372
Brazilian Cystic Fibrosis (CF) physiotherapy care protocol

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The paediatric cystic fibrosis (CF) specialist center at the University Hospital of Campinas Medical school, Brazil, currently has responsibility for the care of 140 children and adolescents with CF. Assessment is a continuous care concern of multi-disciplinary team, which identifies individual problems and needs of the patients and their family. To systematize the dates we elaborated a protocol as a wide investigative tool.
The aim of this protocol is to provide a clinical database of CF patients to facilitate the individual management plans and to identify physiotherapy issues. Methods: This protocol records ten elements: routine physiotherapy techniques, patient pulmonary exacerbation signs and symptoms (from Cystic Fibrosis Foundation), Bhala and Schwaschman scores, growing and body weight assessment, sputum and cough swab culture, lung function tests, cardiac and respiratory parameters evaluation (heart rate, respiratory rate and pulse oximetry), functional ability, postural abnormalities and adherence issues.
Results: this protocol will provided a collection of data for future audit and researches.
Conclusion: This protocol appliance routine, at Brazilian specialist CF center, provides us information about the disease's clinical course and physiotherapy treatment regimen in order to formulate an effective management plan and to elaborate a CF annual physiotherapy review of our patients.

373*
Leg muscle dysfunction in patients with cystic fibrosis (CF) and severe airway obstruction

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Peripheral muscle weakness and atrophy are common findings in patients with CF and severe airway obstruction. The assessment of muscle strength and mass permits to plan an appropriate individualized strength training program. The aims of our study were: 1) to evaluate the relationship between muscle strength and fat free mass (FFM) calculated from skinfolds; 2) to compare quadriceps strength measurement obtained with dynamometer (QS) with leg one-repetition maximal (1RM) tests. 1RM tests are weightlifting exercises which assess strength of different muscle groups. 10 patients with CF (FEV_1 31 ± 6% pred., age 28 ± 7 years) and 10 matched healthy subjects (C) were recruited. Whereas both body mass index and weight were significantly lower in CF patients compared to C, we found no difference in FFM between the two groups. Compared to C, leg muscle strength assessed by QS (14 ± 6 vs 7 ± 4 kg, $p<0.01$) and 1RM tests (squat: 87 ± 30 vs 53 ± 17 kg, $p<0.01$) was significantly reduced in CF patients. There was a significant relationship between both strength measurements and FFM ($p<0.05$) and between QS and 1RM tests ($p<0.05$) in CF patients. In conclusion, the decrease in muscle strength but not in FFM suggests an intrinsic muscle defect in CF patients. 1RM tests are useful to assess limb muscle performance.

374*
Long term effects of positive expiratory pressure (PEP) or oscillatory positive pressure (RC Cornet®) on FEV_1 and perceived health in children with CF

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Aims: This study aimed to examine the long-term effects of the PEP mask (Astra Tech) and the RC Cornet® device on FEV_1 and perceived state of health in children with CF.
Methods: 30 children (20 girls), median age 11 yrs (range 6 – 15) were randomly assigned to use either the PEP mask or RC Cornet® as their primary airway clearance technique (ACT) for 12 months. FEV_1 and perceived state of health were recorded at baseline and 12 months. Children were asked how the device compared to their previous ACT in terms of preference, ease of use and efficacy.
Results: Results from 24 children in this ongoing study have been obtained (14 in PEP). Mean baseline FEV_1 was 75.3% predicted (range 43 – 102) and 63.4 % (range 35 – 95) in the PEP and Cornet groups respectively. After one year there was no significant difference between groups in terms of FEV_1 change, with both groups demonstrating a small improvement (2.2%: PEP, 4.3%: Cornet). Pulmonary exacerbations requiring antibiotics was similar in both groups (8:PEP, 6:Cornet). 13 children perceived their health to be the same at the beginning and end of one year (7 in PEP), while 4 children (3 in PEP) perceived an improvement and 7 (4 in PEP) a reduction. One child in each group stopped using the device as they found it "ineffective and fiddly to clean" (Cornet) or preferred a previously used device (PEP).
Conclusions: Small increases in FEV_1 and predominantly stable perceptions of health were observed in both treatment groups over one year suggesting that both devices were potentially safe and effective in children. Personal preference appears to be a valuable aspect of selecting proven ACT's.

375
Sputum induction (SI) with 3% hypertonic saline in children

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Aim To determine the weight of sputum produced following nebulisation with 3% hypertonic saline and the tolerability of this procedure.
Methods Children 8 years of age or older were recruited. Respiratory function and pulse oximetry were measured before and during the procedure. Children with a history of asthma or routinely take a bronchodilator before physiotherapy were given a bronchodilator before SI. Following mouthwash and gargle, 3% hypertonic saline was delivered by a jet nebuliser using a "Porta-Neb" compressor for 4 three-minute periods. Saliva was collected separately and induced sputum collected after each 3 min cycle. The procedure was stopped following a >20% fall in FEV1 or sustained desaturation (>5% for >1 minute).
Results 13 children with CF were productive of sputum before the procedure (age range, 9.6-17.9 years) and 13 were non-productive (8.2-17.4 years). Sputum weight following SI was significantly greater in the productive children (3.26±2.1g versus 1.42±1.06, $p<0.005$). Children without CF (n=10, age 12.9-17.3 years) produced comparable sputum weights to the non-productive CF children (1.31±0.70g, $p=0.78$). One CF child had a fall in FEV1>20% before the final cycle (despite premedication with salbutamol) and the procedure was stopped at that point. He felt no subjective change. One child had a transient (<1 minute) desaturation of 8% but promptly recovered normal saturations, felt no subjective change in condition and was able to complete the study.
Conclusions Sputum induction with 3% hypertonic saline is well tolerated in children and young people. The weight of sputum was greater in children who were productive but not significantly increased in CF children who were non-productive compared to non-CF controls.